

citoplasmática, tornando as células sanguíneas sensíveis à lise pelo sistema complemento. Além de hemólise intravascular e consequente anemia, a HPN aumenta o risco de trombozes e insuficiência renal. A introdução do eculizumabe como tratamento da HPN de alta atividade melhorou o prognóstico e a sobrevida dos pacientes. O período gestacional, no entanto, devido à ativação do complemento que ocorre sobretudo no terceiro trimestre, cursa com alto risco de eventos tromboembólicos, morte materna, abortamento e parto prematuro. **Conclusão:** Apesar do prognóstico dos pacientes com HPN ter evoluído favoravelmente com o uso de bloqueadores do complemento, a gestação ainda é considerada de alto risco nessa população. O caso relatado ilustra que uma gestante com HPN, mesmo em uso regular de eculizumabe e tromboproliferação, não está livre de um evento desfavorável e ameaçador da vida.

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ALENTUZUMAB AS THIRD LINE TREATMENT IN PURE RED SERIE APLASIA - A SUCCESSFUL CASE REPORT

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Introduction: Pure red cell aplasia (PRCA) is a rare hematologic disorder characterized by reticulocytopenia, normocytic anemia with absence of red cell precursors in bone marrow, normal granulopoiesis, and megakaryopoiesis. The disease may be associated with viral infections, large granular lymphocytic leukemia, chronic lymphocytic leukemia, or thymoma, but the most common form is idiopathic. Refractory PRCA may respond to more profound immunosuppression with anti-CD52. **Purpose:** To describe a refractory PRCA patient treated with anti-CD52 humanized monoclonal antibody, alemtuzumab, with good response. **Case report:** A 52-year-old male patient with hemophilia A, previously treated with cryoprecipitate and currently using prophylactic Factor VIII, presents to healthcare with severe asthenia, no bleeding episodes during the period. Laboratory evaluation: Hb 3.4 g/dL; Ht 10.6% VCM 95; WBC 6.490×10^9 /L; Platelets 196×10^9 /L, intense reticulocytopenia (25×10^9 /L). He had normal renal function, liver function, LDH, vitamin B12, folic acid, iron profile; serology with positive anti-HCV. Bone marrow evaluation showed 50-60% cellularity with intense hypoplasia of the erythroid series, granulocytic series with no changes in morphology or maturation pattern, and normal megakaryocytes with no signs of dysplasia. After investigation with immunoglobulin dosing, chest CT, flow cytometry for LGL, and serology for parvovirus, all normal, the diagnosis of constitutional APSV was confirmed. Treatment with cyclosporine (CSA) at a dose of 6mg/kg was initiated, with no reticulocyte response. In addition, he evolved with progressively increasing bilirubin levels (BT 7.4 BD 5.6) concomitant with a significant increase in Hepatitis C viral load (380,667 IU/mL Log: 5.58).

Immunosuppression was discontinued, treatment with sofosbuvir and simeprevir was initiated, with good viral response. However, he required red blood cell transfusions every 2-3 weeks; new therapy was restarted with CSA and prednisone. The patient did not tolerate this treatment and developed severe thrombocytopenia and no improvement in hemoglobin levels. Considering the high risk of bleeding, CSA was discontinued. Platelet count normalized but he continued to require mensal RBC transfusion. At this point, second-line therapy with IV Alemtuzumab, 3 mg dose followed by 6 weekly 15 mg doses (total 96 mg), coupled with infectious prophylaxis, has been proposed. Response occurred as early as the third infusion, as documented by a rapid increase in reticulocyte count. Within seven weeks the patient had a complete response (Hb 12.4 g/dL, Vg 35.3% and reticulocytes 170×10^9 /L). He had no toxicity-related intercurrents and remained with a complete response after six months of treatment. Unfortunately, a further progressive drop in hemoglobin after nine months from the end of the first treatment was observed. We opted to restart Alemtuzumab at a dose of 15 mg/month, and follow with this maintenance dose. After the third application, the patient again showed a complete response (Hb 14.3 g/dL and VG 40.5%). Currently, the patient continues to use the medication every 45 days with a maintained hemoglobin level. **Conclusion:** Small case series cite Alemtuzumab for refractory APSV as a treatment possibility with 63% overall response. We suggest use in refractory cases, followed by maintenance dosing to avoid relapse in long-term follow-up.

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SEQUESTRO HEPÁTICO EM PACIENTE COM ANEMIA FALCIFORME: UM RELATO DE CASO

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Objetivos: Apresentar um evento raro, em paciente com anemia falciforme, conhecido como: Sequestro Hepático. **Introdução:** A doença da anemia falciforme (AF) frequentemente leva a alterações hepáticas que podem ser classificadas entre agudas e crônicas. Alguns estudos sugerem que a disfunção hepática está mais frequentemente ligada à obstrução crônica de trato biliar ou a hepatite viral, isto é, causas não vasculares (CHARLOTTE, et al. 1985). No entanto, há também estudos indicando que são as alterações vasculares secundárias à doença falciforme desencadeiam lesões hepáticas e, em casos raros, o sequestro hepático (OMATA et al, 1986). **Materiais e métodos:** O relato se baseou em buscas de dados nos prontuários eletrônicos e laudos de exames laboratoriais e de imagem. **Relato de caso:** M.S.D., 40 anos, sexo feminino, dá entrada no serviço de emergência informando ser portadora de anemia falciforme, com astenia intensa, piora da icterícia, anúria e dor no quadrante superior direito do abdômen. Ao exame físico apresentava-se dispnéia, taquicárdica, com edema bilateral em membros inferiores (4+/4+), icterícia (2+/4+), ritmo cardíaco regular em 2T, com bulhas normofonéticas, sem sopros, murmúrios vesiculares